Complete Summary

GUIDELINE TITLE

Diagnosis and treatment of childhood hypercholesterolaemia.

BIBLIOGRAPHIC SOURCE(S)

Finnish Medical Society Duodecim. Diagnosis and treatment of childhood hypercholesterolaemia. In: EBM Guidelines. Evidence-Based Medicine [Internet]. Helsinki, Finland: Wiley Interscience. John Wiley & Sons; 2007 Feb 13 [Various].

GUIDELINE STATUS

This is the current release of the guideline.

This guideline updates a previous version: Finnish Medical Society Duodecim. Diagnosis and treatment of childhood hypercholesterolaemia. In: EBM Guidelines. Evidence-Based Medicine [CD-ROM]. Helsinki, Finland: Duodecim Medical Publications Ltd; 2005 Mar 21. various p.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

Hypercholesterolemia

GUIDELINE CATEGORY

Diagnosis
Evaluation
Management
Prevention

Screening
Treatment

CLINICAL SPECIALTY

Endocrinology
Family Practice
Pediatrics
Preventive Medicine

INTENDED USERS

Dietitians
Health Care Providers
Physicians

GUIDELINE OBJECTIVE(S)

Evidence-Based Medicine Guidelines collect, summarize, and update the core clinical knowledge essential in general practice. The guidelines also describe the scientific evidence underlying the given recommendations.

TARGET POPULATION

Children (>2 years of age) who may be at risk for hypercholesterolemia or who are diagnosed with hypercholesterolemia

INTERVENTIONS AND PRACTICES CONSIDERED

Prevention/Screening/Evaluation/Diagnosis

1. Screening for hypercholesterolemia on basis of family history
    - Measurement of fasting serum cholesterol, high-density lipoprotein (HDL) cholesterol, and triglycerides
    - Calculation of low-density lipoprotein (LDL) cholesterol (Friedewald's formula)
2. Double-checking of increased values
3. Exclusion of secondary hyperlipidemias by measuring serum free T_4, serum thyroid stimulating hormone (TSH), serum alanine aminotransferase (ALT), and urine albumin
4. Patient education and consultation at a genetic unit, as indicated

Treatment/Management

1. Diet (decreased saturated fat) with follow-up at appropriate intervals (e.g., 3, 6, and 12 months)
2. Regular physical training and active everyday exercise
3. Referral to specialist, as indicated (pediatric endocrinologist, dietician, pediatric clinic)
4. Drug therapy (resin, statin) as indicated

MAJOR OUTCOMES CONSIDERED

- Serum cholesterol levels
- Safety of treatment interventions

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Primary Sources)
Hand-searches of Published Literature (Secondary Sources)
Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

The evidence reviewed was collected from the Cochrane database of systematic reviews and the Database of Abstracts of Reviews of Effectiveness (DARE). In addition, the Cochrane Library and medical journals were searched specifically for original publications.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence

A. **Quality of Evidence: High**

Further research is very unlikely to change confidence in the estimate of effect

- Several high-quality studies with consistent results
- In special cases: one large, high-quality multi-centre trial

B. **Quality of Evidence: Moderate**

Further research is likely to have an important impact on confidence in the estimate of effect and may change the estimate.

- One high-quality study
- Several studies with some limitations

C. **Quality of Evidence: Low**

Further research is very likely to have an important impact on confidence in the estimate of effect and is likely to change the estimate.

- One or more studies with severe limitations

D. **Quality of Evidence: Very Low**

Any estimate of effect is very uncertain.

- Expert opinion
- No direct research evidence
- One or more studies with very severe limitations

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Not stated

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

The levels of evidence [A-D] supporting the recommendations are defined at the end of the "Major Recommendations" field.

Aim

- To identify children with hypercholesterolaemia on the basis of a family history (parents) of coronary heart disease and high blood lipid levels. Screening the entire child population is not recommended.

Directing Screening at Risk Families

- Hypercholesterolaemia should be searched for in families with precocious coronary heart disease.
    - Father or grandfather at age <55 years, or mother or grandmother at age <65 years, or
    - Hyperlipidaemia
        - Serum cholesterol $\geq$8.0 mmol/L or
        - Serum low-density lipoprotein (LDL) cholesterol $\geq$6.0 mmol/L or
        - Serum triglycerides $\geq$5.0 mmol/L or
        - Milder hyperlipidaemia with low (<0.9 mmol/L) serum high-density lipoprotein (HDL) cholesterol
- At screening every family member older than 2 years of age should have their fasting serum cholesterol, HDL cholesterol, and triglycerides measured after a 12-hour fast, and LDL cholesterol calculated with Friedewald's formula. (See Finnish Medical Society Duodecim guideline "Lipid Measurements and Their Sources of Error: LDL Cholesterol.")
- Increased values measured at screening should be double-checked.
- Secondary hyperlipidaemias should be excluded by measuring serum free T_4, serum thyroid stimulating hormone (TSH), serum alanine aminotransferase (ALT), and urine albumin before commencing therapy.
- Young patients with coronary heart disease and hyperlipidaemias should be informed of the importance of having their children and grandchildren examined. A general practitioner or an internist may initiate directed screening. Departments of internal medicine and paediatrics should agree on examination of the children and coordinate assessment of results.
- If the family history indicates frequent coronary heart disease, the finding of hyperlipidaemia may be a cause of anxiety for the nearest relatives. As accurate evaluation of the family history often requires informing and assessing persons living in various parts of the country, a consultation at a genetic unit can be considered.

Classification of Hypercholesterolaemia

Table. Classification of Hypercholesterolaemias in Childhood

	Serum Cholesterol (mmol/L)	Serum LDL Cholesterol (mmol/L)
Not increased	<5.5	<4.0

Table. Classification of Hypercholesterolaemias in Childhood

	Serum Cholesterol (mmol/L)	Serum LDL Cholesterol (mmol/L)
Increased	5.5-6.9	4.0-5.4
Significantly increased	≥7.0	≥5.5

Therapy: Indications and Practice

- Serum cholesterol of less than 5.5 mmol/L (LDL <4.0) does not require further action. In borderline cases, the general dietary advice is given or repeated.
- With an increased serum cholesterol, it usually suffices to commence a diet and follow up the child at 3, 6, and 12 months. If a diet maintained for 6 to 12 months does not decrease serum cholesterol to below 5.5 mmol/L or LDL cholesterol below 4.0 mmol/L, the child should be remitted to a paediatric clinic for assessment by a paediatric endocrinologist or a paediatrician familiar with therapy of hyperlipidaemias. If necessary, a dietician should be used for dietary instructions. The child should be motivated to regular physical training and active everyday exercise, which have a beneficial effect on hyperlipidaemia.
- A child with significantly increased serum cholesterol should be remitted directly to a paediatric clinic.
- The need for drug therapy is decided mainly on family history of coronary heart disease. Drug therapy (a resin is the first-line drug [Tonstad et al., 1996; West, Lloyd, & Leonard, 1980; Glueck et al., 1986] [**B**]; a statin may be used as an alternative) is initiated by an experienced paediatrician.
- Drug therapy is rarely needed before puberty, and very rarely before school age.

Diet

- Diet is the single most important treatment for hyperlipidaemia, and it may be sufficient even for familial hypercholesterolaemia in childhood. The diet should be followed from the age of two years. It is most important to decrease the amount of saturated fat.
    - Reduction in the use of dairy fat
        - Skim milk or 1% fat milk
        - No- or low-fat dairy products and cheeses
    - Sitostanol- and sitosterol-containing margarine or vegetable oil-based margarine on bread
    - Reduction in the use of fatty veal or pork
    - Use of fibre-rich and full corn products, oatmeal, and fish is encouraged.
    - To maintain adequate calcium intake, total abstention from dairy products is not recommended.

Related Resources

- Sitostanol margarine appears to decrease serum cholesterol in children with familial hypercholesterolaemia (Gylling, Siimes, & Miettinen, 1995) [**C**].
- Two years of pravastatin therapy appear to induce regression of carotid atherosclerosis in children with familial hypercholesterolemia, without significant adverse effects (Wiegman et al., 2004) [**B**].

Definitions:

Levels of Evidence

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CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

REFERENCES SUPPORTING THE RECOMMENDATIONS

References open in a new window

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

Concise summaries of scientific evidence attached to the individual guidelines are the unique feature of the Evidence-Based Medicine Guidelines. The evidence summaries allow the clinician to judge how well-founded the treatment recommendations are. The type of supporting evidence is identified and graded for select recommendations (see the "Major Recommendations" field).

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

- Improved identification of children at risk for development of hypercholesterolemia
- Appropriate diagnosis and treatment of childhood hypercholesterolemia

POTENTIAL HARMS

Not stated

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness
Staying Healthy

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

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ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2004 Jun 14 (revised 2007 Feb 13)

GUIDELINE DEVELOPER(S)

Finnish Medical Society Duodecim - Professional Association

SOURCE(S) OF FUNDING

Finnish Medical Society Duodecim

GUIDELINE COMMITTEE

Editorial Team of EBM Guidelines

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Primary Author: Matti Salo

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

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GUIDELINE AVAILABILITY

This guideline is included in a CD-ROM titled "EBM Guidelines. Evidence-Based Medicine" available from Duodecim Medical Publications, Ltd, PO Box 713, 00101 Helsinki, Finland; e-mail: info@ebm-guidelines.com; Web site: www.ebm-guidelines.com.

AVAILABILITY OF COMPANION DOCUMENTS

None available

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI on August 30, 2005. This summary was updated by ECRI on October 26, 2005. This NGC summary was updated by ECRI Institute on November 13, 2007.

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Date Modified: 9/22/2008


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